Case Reports

Intermittent Cyanosis Years after a Mustard Repair

for Dextro-Transposition of the Great Arteries

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A 28-year-old woman, who as an infant had undergone the Mustard atrial switch procedure for dextro-transposition of the great arteries, presented with a baffle leak and consequent intermittent cyanosis. In addition, an occlusive thrombus had formed in the systemic venous baffle after a failed attempt to remove infected pacemaker leads. Corrective surgery was successful. In addition to the case of our patient, we discuss long-term sequelae of the atrial switch procedure that present challenges in patient care. ***(Tex Heart Inst J 2012;39(5):665-7)***

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Dextro-transposition of the great arteries (D-TGA) is a cyanotic congenital heart defect wherein the main pulmonary artery and aorta are transposed: the pulmonary artery arises from the left ventricle and the aorta arises from the right ventricle.[1] Consequently, oxygenated blood is restricted to the pulmonary circulation, and deoxygenated blood remains in the systemic circulation, resulting in 2 separate circulations in parallel. Profound cyanosis results unless there is a major intracardiac shunt or patent ductus arteriosus. Most D-TGA patients who have survived to adulthood have undergone an early surgical correction called the atrial switch procedure, which was first described by Andersen and Senning in 1959[2] and then by Mustard and colleagues in 1964.[3] This operation (also called a Senning or Mustard procedure) redirects the systemic venous blood from the venae cavae to the subpulmonic left ventricle through an intra-atrial baffle. Oxygenated blood then returns from the pulmonary veins into the systemic right ventricle through a pulmonary venous baffle. Many atrial-switch patients are now reaching their 3rd or 4th decade of life, and sequelae related to this procedure continue to present challenges. We present the case of a woman who underwent the Mustard procedure as an infant and presented with intermittent cyanosis as an adult.

Case Report

A 28-year-old woman had undergone a Mustard operation for D-TGA repair at 6 months of age. Postoperative sinus node dysfunction developed, and an epicardial intra-abdominal pacemaker was implanted. This was later converted to a transvenous pacing system. The patient did well throughout childhood and adolescence. At age 28, she was admitted to her local hospital because of recurrent infections in the subclavicular pacemaker pocket site after a pulse generator replacement. During this hospitalization, she was treated for septic shock, respiratory distress, and intermittent cyanosis. The pulse generator was thought to be the source of infection, so it was removed. However, an attempt to extract the leads was unsuccessful. In addition, an echocardiogram suggested an inferior baffle leak.

The patient was transferred to the Adult Congenital Heart Disease Program at Texas Children's Hospital. Physical examination showed a heart rate of 63 beats/min, a blood pressure of 112/55 mmHg, and a temperature of 97.1 °F. The patient's pulse oximetry reading was 79% on 6 L of oxygen, with frequent desaturation to 40% with no apparent precipitating factors. A grade 3/6 systolic ejection murmur was heard at the left sternal border, together with an S_3 and a right ventricular heave. The patient was noticeably cyanotic, with blue nail beds but no clubbing. A transesophageal echocardiogram showed a baffle leak in the inferior vena cava graft; in addition,

a sessile mass attached to the transvenous ventricular pacemaker lead intermittently obstructed the superior portion of the systemic venous baffle from entering the subpulmonic left ventricle (Fig. 1). Angiography confirmed the systemic venous baffle leak into the pulmonary venous baffle (Fig. 2), which created a right-to-left intracardiac shunt. Cardiac catheterization revealed an oxygen saturation of 60% in the aorta and a calculated Qp/Qs ratio of 0.5:1. Superior vena cava (SVC) pressures were elevated at 20 mmHg.

Blood cultures grew coagulase-negative staphylococcus bacteria, so the patient underwent 6 weeks of treatment with intravenous vancomycin. She was referred for surgical repair of the baffle leak and removal of the mass on the transvenous ventricular pacemaker lead. A bovine pericardial patch was placed in the right lateral SVC-to-inferior vena cava wall to correct the shunt (Fig. 3), the mass was excised, additional epicardial pacing wires were placed, and the transvenous pacing leads were removed. The patient was discharged from the hospital on postoperative day 6, with atrioventricular sequential pacing. She had an oxygen saturation level of 95% on room air. At her 1-year follow-up examination, her oxygen saturation remained acceptable, her pacemaker function was normal, and she was in New York Heart Association functional class II.

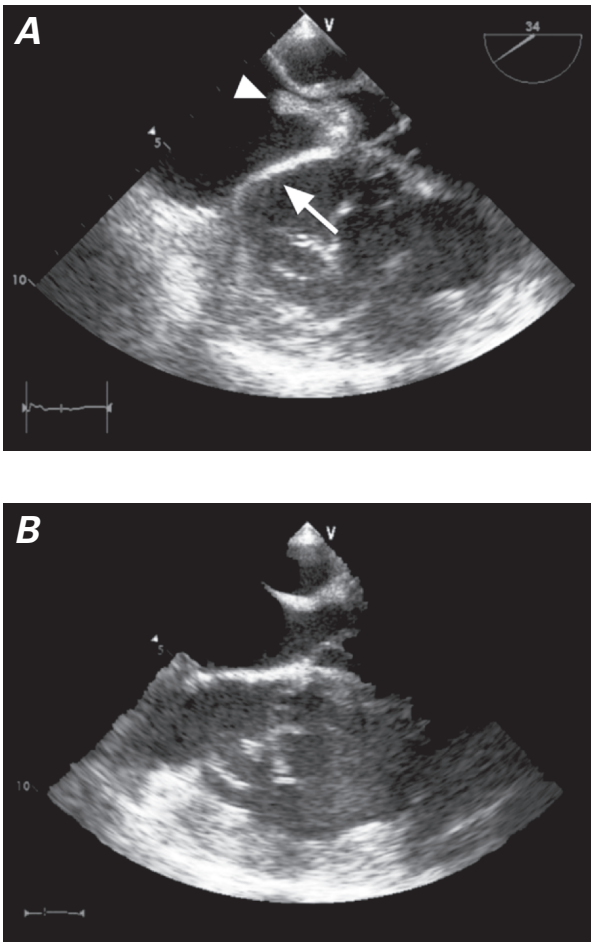

Fig. 1 *Transesophageal echocardiograms.* ***A****) A mass (arrowhead) obstructs the venous return of the systemic venous baffle (arrow).* ***B****) Appearance after removal of the mass.*

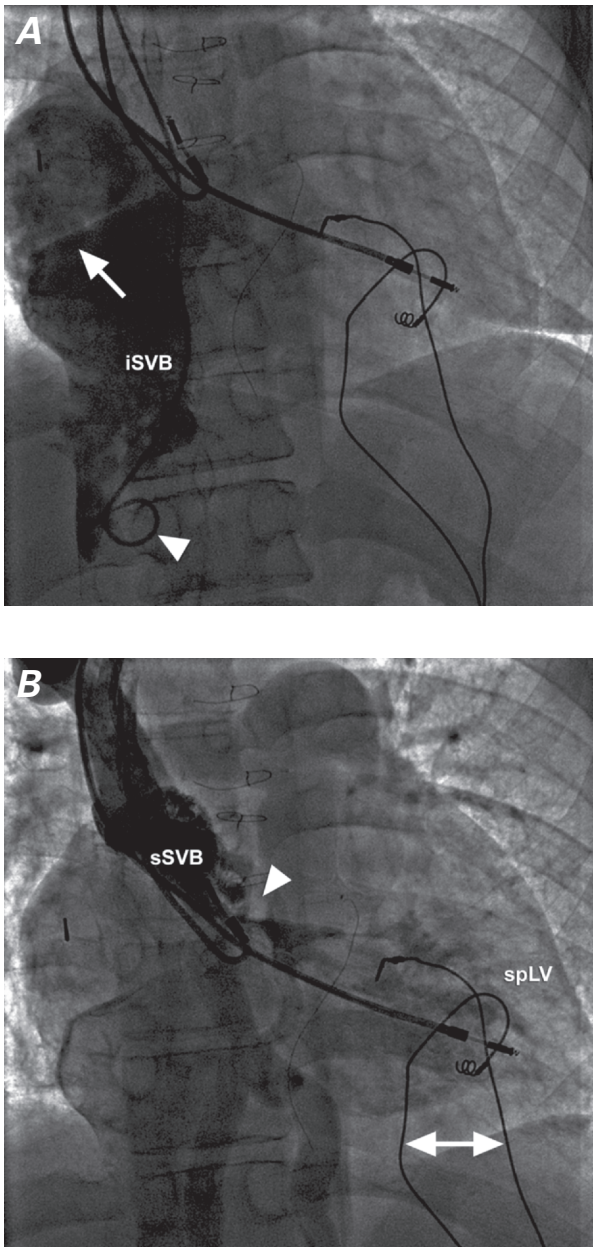

Fig. 2 *Coronary angiograms (anteroposterior projection).* ***A****) Angiogram shows a pigtail catheter in the inferior vena cava (arrowhead), return of contrast medium into the inferior portion of the systemic venous baffle (iSVB), and leakage of contrast medium across the atrial baffle. Arrow shows the linear demarcation between the systemic and pulmonary venous baffles.* ***B****) The superior portion of the systemic venous baffle (sSVB) shows near-occlusion (arrowhead) of venous return to the subpulmonic left ventricle (spLV). Double arrow shows the transvenous pacemaker lead in the spLV and the abandoned epicardial pacemaker wires.*

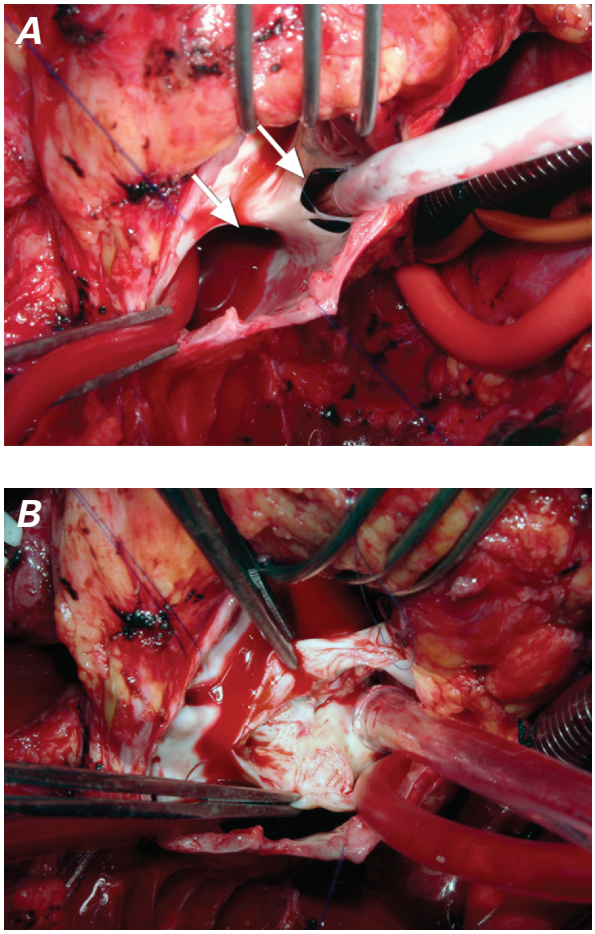

Fig. 3 *Intraoperative photographs show* ***A****) the atrial baffle leaks before repair (white arrows) and* ***B****) the surgical closure with a pericardial patch.*

Discussion

Decades after the atrial switch procedure was introduced, long-term sequelae continue to present challenges in patient care. Superior vena cava obstruction and baffle leaks are long-term sequelae that have been previously reported.[4] Together, they account for approximately 60.2% of all corrective procedures in patients who once underwent the atrial switch repair.[5,6] Superior vena cava obstruction occurs in 5% to 10% of patients who have undergone the Mustard procedure, and intervention has been required in approximately 4% of these patients.[7,8] Atrial baffle leaks occur more frequently than SVC complications; however, only 1% to 2% of baffle leaks produce symptoms that are significant enough to warrant surgical intervention.[7,9]

Arrhythmias are very common and increase with age after atrial switch procedures. Sinus node dysfunction (28% at 5 yr and 39% at 10 yr postoperatively)[5,10] can lead to sick sinus syndrome. Intra-atrial reentrant tachycardia, atrial tachycardia, or atrioventricular node reentry—common arrhythmias in this patient population—are often caused by a tachycardia-bradycardia syndrome. Studies have shown that atrial flutter and intra-atrial reentrant tachycardia are predisposing markers for sudden death,[2,11] and 11% to 38% of patients have had pacemakers implanted as therapy for these arrhythmias.[2,11] As these atrial-switch patients age, sequelae related to pacemakers (such as infection and thrombus formation) necessitate lifelong monitoring. The prevalence of device-related infections is as high as 3.2% for implantable cardioverter-defibrillators and 19.9% for pacemakers in these patients.[12] Our patient experienced several of the sequelae that can occur in an adult congenital heart disease patient with D-TGA who has undergone an atrial switch procedure in childhood.

References

1. Warnes CA. Transposition of the great arteries. Circulation 2006;114(24):2699-709.
2. Andersen MN, Senning A. A study of reinforced ivalon as prosthetic material for arterial replacement. Arch Surg 1959; 79:542-7.
3. Mustard WT, Keith JD, Trusler GA, Fowler R, Kidd L. The surgical management of transposition of the great vessels. J Thorac Cardiovasc Surg 1964;48:953-8.
4. Dragulescu A, Sidibe N, Aubert F, Fraisse A. Successful use of covered stent to treat superior systemic baffle obstruction and leak after atrial switch procedure. Pediatr Cardiol 2008;29(5): 954-6.
5. Murphy DJ Jr. Transposition of the great arteries: long-term outcome and current management. Curr Cardiol Rep 2005;7 (4):299-304.
6. Oechslin E, Jenni R. 40 years after the first atrial switch procedure in patients with transposition of the great arteries: long-term results in Toronto and Zurich. Thorac Cardiovasc Surg 2000;48(4):233-7.
7. Lange R, Horer J, Kostolny M, Cleuziou J, Vogt M, Busch R, et al. Presence of a ventricular septal defect and the Mustard operation are risk factors for late mortality after the atrial switch operation: thirty years of follow-up in 417 patients at a single center. Circulation 2006;114(18):1905-13.
8. Sharaf E, Waight DJ, Hijazi ZM. Simultaneous transcatheter occlusion of two atrial baffle leaks and stent implantation for SVC obstruction in a patient after Mustard repair. Catheter Cardiovasc Interv 2001;54(1):72-6.
9. Kirklin JW, Colvin EV, McConnell ME, Bargeron LM Jr. Complete transposition of the great arteries: treatment in the current era. Pediatr Clin North Am 1990;37(1):171-7.
10. Cobanoglu A, Abbruzzese PA, Freimanis I, Garcia CE, Grunkemeier G, Starr A. Pericardial baffle complications following the Mustard operation. Age-related incidence and ease of management. J Thorac Cardiovasc Surg 1984;87(3):371-8.
11. Kammeraad JA, van Deurzen CH, Sreeram N, Bink-Boelkens MT, Ottenkamp J, Helbing WA, et al. Predictors of sudden cardiac death after Mustard or Senning repair for transposition of the great arteries. J Am Coll Cardiol 2004;44(5):1095-102.
12. Cesario D, Kedia R, Desai N, Aboulhosn J, Uslan D, Boyle N, et al. Device extraction in adults with congenital heart disease. Pacing Clin Electrophysiol 2009;32(3):340-5.